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Urinary Diversion and Undiversion

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Introduction

The current practice of urinary diversion and undiversion in children has a long history of trial and error. In this article we trace the evolution of current views on the subject.

Urinary Diversion

The majority of pediatric urinary diversions are carried out for management of congenital anomalies. Other reasons for diversion include trauma, malignancy, neurologic disorders, severe inflammatory bladder disease and failed previous surgery.[1,2] Over the past 30 years the incidence of urinary diversions has been decreasing, not only because better methods of managing these children have developed but also because it has become apparent that urinary diversions have complications of their own and often diversion does not serve its intended purpose.

Urinary diversion in children dates back to 1852 when Sir John Simon diverted a patient with bladder exstrophy.[3,4] However, it was not until the ileal loop was popularized[5] that a reliable, reproducible and generally applicable method of urinary diversion became available. Ileal conduits were used exclusively for many years until it was recognized that they produce unique problems in children. Since then various methods have been devised to circumvent these problems.

The primary goal of urinary diversion is to protect the urinary tract proximal to the problem area. Typically urinary diversions are classified as permanent or temporary. Historically, permanent diversions were carried out in children with severe incontinence (bladder exstrophy, neurogenic bladder, ectopic ureter), progressive upper tract deterioration [posterior urethral valves, neurogenic bladder, prune belly syndrome, severe vesico-ureteric reflux (VUR)] or ablative surgery for pelvic tumors (rhabdomyosarcoma of the bladder, prostate, or female genitalia). Today these children are only considered for permanent diversion when corrective procedures have unquestionably failed.

Temporary diversions were often carried out in the past for conditions that should have been corrected primarily (*e.g.* uretero-pelvic junction (UPJ) obstruction and severe VUR). Unfortunately, at times, diversion resulted in infections and secondary damage to these kidneys effectively precluding corrective surgery.

Currently temporary diversions are indicated for the following purposes:

1. Emergently address metabolic abnormalities or sepsis
2. Preserve renal function until more definitive measures can be taken
3. Allow time for growth so subsequent reconstruction can be accomplished more safely[6]

Temporary Diversion

Temporary diversion may be carried out with or without an indwelling tube or stent.

Intubated Temporary Diversion

Nephrostomy

Nowadays open nephrostomies are only carried out if pelvicalyceal injury is noted upon exploration for trauma. In all other circumstances interventional radiologists place nephrostomy tubes under ultrasonographic and/or fluoroscopic guidance. The percutaneous technique was first described by Goodwin.[7] Due to its simplicity it is used frequently as a temporizing maneuver in children with acute renal obstruction and in the chronically obstructed poorly functioning kidney. In the latter case, preoperative drainage helps evaluate function of the affected kidney, thus helping us to choose whether to reconstruct the system or to carry out a nephrectomy.

In intubated nephrostomies inadvertent dislodgement of the tube is very common. Calculus formation, leak of urine around the tube and chronic infection also occur frequently.[8] Chronic infection eventually leads to progressive renal failure.[6] In addition, UPJ obstruction has been reported in infants after prolonged nephrostomy drainage.[9] In percutaneously placed nephrostomies perirenal hematoma and perinephric urinoma are also known to have formed.[10] In addition the procedure carries risks inherent in blind procedures namely hemorrhage and injury to adjacent structures such as intestines and pleura.[11]

Generally a straight tube with a self-retaining mechanism shaped like a mushroom, pigtail or "J" is used. Occasionally a circle or loop nephrostomy tube with multiple side holes is placed. Obviously such a tube will not fall out but the holes should be placed accurately so as to lie within the collecting system. A circle nephrostomy is particularly useful if a cutaneous pyelostomy cannot be carried out either because the renal pelvis is intrarenal or it is not dilated.[12,13,14]

Ureteral Stents and Intubation

The end/s of stents could be brought out of the body or be totally internal. Externalized stents are rarely used since they tend to get blocked, kinked and dislodged. If used they are generally inserted antegrade after a percutaneous nephrostomy. When ureters are intubated for a short period, such as after ureteric reconstruction the distal end of the tube is often brought out at the abdominal wall for ease of removal. Externalized stents present a route for organisms to enter the urinary tract. Internal stents are often used for temporary drainage of an obstructed system and to protect suture lines, prevent leakage, and to immobilize and align the ureter after repair or reconstruction of the urinary tract. Even patients with internal stents are at a higher risk for infections as the stent constitutes a foreign body and concretions develop on it in due course of time. Short-term use of antibiotics or antibiotic irrigation are both valuable adjuncts in preventing infections. In small caliber ureters of children, width and length of the stent must be carefully measured to prevent pressure necrosis of the ureteric wall and pressure by the ends of the stent on the kidney at one end and the bladder wall at the other. Stents can be placed antegrade through the kidney or retrograde via a cystoscope. The lower end of the stent can be brought out at the urethra but in most instances both ends are internalized and cystoscopy is required for their removal. Stents retain their position because they have a pig-tail configuration at both ends.

Cystostomy

Open cystostomy is usually carried out as part of another procedure such as removal of a large bladder calculus or repair of the bladder and rarely for urinary retention. In children it is also preferred, over the percutaneous method, if they have had previous lower abdominal or pelvic surgery since postoperative adhesions not only distort the anatomy but intestinal loops may be adherent to the anterior wall of the bladder. Cystostomy is to be avoided even in children with infravesical obstruction if they have poor ureteral peristalsis, secondary ureteral dilation and tortuosity since it does not improve drainage from the kidneys and it may exacerbate obstruction at the ureterovesical junction.[15,8] Specific complications consist of stone formation, infection, retraction of the anterior bladder wall and bladder spasms due to irritation of the trigone by the tip of the catheter. Apart from the exceptional circumstances mentioned earlier, we prefer the percutaneous technique for patients in urinary retention. In the percutaneous method, in addition to complications mentioned for the open technique, inadvertent bowel injury is possible hence the catheter is best placed under ultrasound guidance.

Perineal Urethrostomy

Intubated perineal urethrostomies are very rare today. In the past they were used for urinary diversion after the second stage of a proximal hypospadias repair, instead of a suprapubic cystostomy. In our experience, bladder spasms occur less often with an intubated perineal urethrostomy when compared to a suprapubic catheter resting against the trigone. Apart from catheter dislodgement, a technical complication is bleeding from the corpus spongiosum if one deviates from the midline or if multiple incisions are made to locate the urethra. Urethrocutaneous fistula and stricture could occur once the urethrostomy closes.

Urethral Catheterization

Simple urethral catheterization fulfills all the criteria of intubated urinary diversion. The most dramatic example of its value is in the patient with posterior urethral valves. A bladder catheter placed transurethrally allows time to correct azotemia and electrolyte abnormalities. It also permits us to evaluate renal functions and to determine whether aperistaltic ureters are contributing to renal compromise. In children with posterior urethral valves we find it easier to insert a feeding tube of appropriate size rather than a soft Foley catheter, which tends to curl up in the dilated posterior urethra. In addition, the balloon of the Foley catheter may compress the ureteric orifices thus compromising drainage instead of improving it.[16]

Tubeless Temporary Diversion

Cutaneous Pyelostomy

Exteriorizing the renal pelvis to the skin permits the kidney to drain directly and since there are no tubes, foreign body reaction and dislodgement do not occur. In addition, unlike the ureter, the very vascular renal pelvis is not rendered ischemic either during initial diversion or at subsequent closure. Known complications are skin irritation, appliance failure and stomal stenosis.[6] While it is not a problem for the infant in diapers the inability to effectively apply an appliance is a particularly distressing problem for the older child. In case of a large flabby kidney the stoma should be of the smallest size that permits adequate drainage since the renal pelvis and even the kidney may prolapse. The procedure can only be performed if the patient has a large extrarenal pelvis.[17,18]

Cutaneous Ureterostomy

The popularity of ureterostomies has waned in the past three decades because pyelostomies offer much better drainage. Often, the lack of peristalsis in these dilated ureters results in stasis which, in turn, increases the chances of urinary infection. Furthermore, stomal stenosis is not uncommon, particularly if normal sized ureters are exteriorized.[19,20] Finally, at the time of takedown or subsequent undiversion, the distal ureter can be devascularized if its longitudinal blood supply is damaged.

Ureterostomies are fundamentally of three types: Loop ureterostomy which was first described by F. Douglas Stephens in 1960.[21] It is technically the easiest to perform and it was used almost exclusively to temporize in children with massively dilated ureters secondary to posterior urethral valves. It has fallen out of favor since these large aperistaltic ureters do not always drain as well as anticipated and high complete diversion defunctionalizes the bladder. Defunctionalizing the bladder was considered deleterious to eventual bladder growth and function.[22] This belief is now being questioned and Jayanthi *et al*[23] are of the opinion that poor bladder compliance in patients with posterior urethral valves is an inherent and unavoidable developmental problem. A variant of the loop ureterostomy is the ring ureterostomy in which the afferent and efferent limbs are anastomosed. This permits urine to flow distally into the bladder.[24] Retik and Perlmutter[8] have suggested that closure of a loop ureterostomy should be delayed for many months after lower ureteral surgery to avoid devascularising the distal ureter. Others have suggested that if care is taken to preserve the blood supply, both, the upper and lower ureter can be operated upon at the same time.[25,26] The Sober Y-ureterostomy is a variation of the loop ureterostomy that permits some urine to pass into the distal ureter and bladder thus, in theory, the bladder is not totally defunctionalized. It is also the easiest to close, with minimal risk of distal ureteral devascularization.[27,28,29] Finally there is the end ureterostomy. The ureters may be brought out either as two separate stomas or as a single cutaneous stoma after constructing a transureteroureterostomy (TUU). Retik and Perlmutter[8] have suggested its use in patients with end-stage renal disease prior to transplantation. In such situations it should be considered for permanent diversion. For a TUU it is important to make the retroperitoneal tunnel wide enough that the crossing ureter is not compressed, and that it has a smooth curve to prevent kinking. The tunnel must be kept clear of the inferior mesenteric artery. Finally, before constructing a TUU one must bear in mind how it would affect the subsequent undiversion. Since stomal stenosis is not uncommon with end ureterostomies, skin flaps and Z-plasties have been recommended.[30]

Cutaneous Vesicostomy

Cutaneous vesicostomy is a simple and highly effective method of diverting the obstructed or dysfunctional lower urinary tract. It has been used for children with posterior urethral valves (PUV), in whom valve ablation is not immediately possible for technical reasons,[31,32] in patients with neurogenic bladders where clean intermittent catheterization (CIC)[33] is not feasible,[34,35,36] and even in some patients with severe VUR who are awaiting ureteric reimplantation.[20,32,8] Duckett[37] first popularized the use of vesicostomies in patients with PUV. In addition to affording excellent drainage from the bladder and preventing high intravesical pressures, since the vesicostomy is carried out in a nondependent location, it does not defunctionalize the bladder. To ensure

adequate renal drainage in patients with infravesical obstruction, it is necessary to identify patients who also have concomitant ureterovesical junction obstructions, prior to carrying out a vesicostomy. A Glucoheptonate or Mag 3 diuretic renal scan with a draining bladder catheter in place will usually provide this information. A Whitaker pressure study could also be carried out[38] but it is usually not required. In children, since the bladder is an abdominal organ, the Blocksom vesicostomy[39] works very well and it is easy to perform and to reverse. Apart from prolapse of the bladder wall other complications that occur, particularly with prolonged usage, are incomplete bladder emptying, bacterial colonization and formation of vesical calculi.[40,41,35] Lapides[42] described a technique of vesicostomy involving skin and detrusor flaps so as to prevent stomal stenosis. This procedure is complex and not really necessary if the stoma is matured properly hence it never became popular. Cutaneous vesicostomies also produce a social problem, particularly in the pubertal child, since it is difficult to fit an appliance once pubic hair develops.

Perineal Urethrostomy

A tubeless perineal urethrostomy is rarely if ever used today. In the past it was occasionally used in males who required CIC but had difficulty catheterizing due to formation of false passages and strictures or because they had a sensate urethra.[43] When the urethrostomy is no longer required, a formal urethroplasty, possibly with a skin patch, is required for closure. Stomal stenosis and ascending infection to the bladder are complications to be watched for. After closure of the urethrostomy a urethrocutaneous fistula or stricture can develop.

Permanent Diversion

Permanent urinary diversion is now rather uncommon in children for the following reasons:

1. The introduction of Clean Intermittent Catheterization by Lapides,[42] which revolutionized our concepts regarding bladder emptying and resulted in the development of reservoirs that do not have to empty spontaneously.
2. The seminal contributions of Hendren[44,45] who, against conventional wisdom of the time, reconstructed children that would normally have been diverted. He then went a step further and undiverted many children who had "permanent" diversions. In both groups he demonstrated that the children did well if they were cared for properly.
3. The introduction of urodynamic evaluations which allowed better assessment of bladder (and ureteric) function and introduced a more selective approach to functional problems.
4. Improved understanding of the physiology of the infant and child which led to major advances in pediatric anesthesiology and intensive care. These advances permitted surgeons to safely carry out complex, long drawn out and demanding operations.

The few children who are permanently diverted nowadays consist of patients with tumors of the lower urinary tract, some patients with bladder or cloacal exstrophy who are not amenable to reconstruction, and quadriplegic and institutionalized children. Very occasionally patients with severely dilated upper urinary tracts, who will not get adequate care at home, may also be candidates for permanent diversion.

Permanent diversion must only be considered after all other treatment options have been exhausted. It should also be remembered that refluxing conduits hamper normal renal growth in the child.[46] In addition stomas and appliances could create devastating psychosocial problems in the adolescent.[47]

In permanent diversions the ureters are anastomosed to a gastrointestinal (G.I.) segment, usually small or large bowel. Diversions can be external or internal. External diversions are either non-continent (appliance dependent) or continent and internal diversions could drain ectopically (into colon) or they may be orthotopic (substitution cystoplasty). Finally, the ureters can be implanted into the conduit so as to permit or prevent reflux.

It bears reiterating that while performing intestinal anastomoses, the following basic guidelines must be followed. The patient's bowel must be prepared prior to surgery and the ends of severed bowel must be occluded with atraumatic clamps to prevent spill into the wound. The abdominal incision must be large enough to provide excellent exposure without straining. Blood supply to the severed ends of bowel must not be compromised and a tension-free and accurate apposition of serosa to serosa must be obtained without strangulation of the tissues. Finally, mesenteric gaps must be closed to prevent internal herniation.[48]

Uretero-intestinal Anastomosis

To ensure unobstructed drainage into the conduit there must not be any sharp curves in the ureter or at the anastomosis. To prevent stenosis at the anastomosis, the ureter must have a good blood supply, there should be no tension at the anastomotic site and accurate mucosa to mucosa approximation of the ureter with the intestine is essential. The anastomosis could reflux freely or be nonrefluxing.

Refluxing Anastomoses

Refluxing uretero-intestinal anastomoses are infrequently performed in children because of complications associated with intestino-ureteric-renal reflux.

Non-refluxing Anastomoses

A non-refluxing ureter to small bowel anastomosis was described by LeDuc[48] but it is a complicated procedure which has not gained popularity. A non-refluxing anastomosis is easier to make in large intestine where the ureter can be placed in a submucosal tunnel.[49] Leadbetter[50] combined Coffey's technique with direct mucosal anastomosis in an effort to prevent reflux and obstruction. Goodwin and his associates obtained the same results by an open approach a few years later.[51]

Non-continent Diversion

Jejunal Conduit

The use of jejunum for urinary diversion never gained favor since it is technically more challenging than any other intestinal conduit and hyponatremia, hyperkalemia and acidosis are common.[52] At present the only possible indications for using jejunum is when ileum and colon are damaged either by pelvic radiation or inflammatory bowel disease or if one wishes to create a large pyelo-enteric diversion for easy passage of renal stones. The proximal end of the segment can be anastomosed to a single kidney. If bilateral drainage is desired, the proximal end of the intestine is anastomosed to one renal pelvis. The intestine is then run past the contralateral renal pelvis which is anastomosed to the side of the conduit. Both pelves can also be drained by employing two smaller segments of intestine, the proximal ends of which are anastomosed to a renal pelvis each and the distal ends to each other in a Roux-en-Y fashio.[53]

Ileal Conduit

The ileal conduit became popular once severe electrolyte imbalances created by ureterosigmoidostomies became evident.[54] Tizzoni and Foggi[55] were the first to describe the procedure but it was left to Bricker[5] to popularize it. For many years the ileal conduit was the most commonly used method of urinary diversion. In part, its popularity was based on the ease with which it can be formed. The long mesentery permits the external end to be brought out without tension and an appliance can easily be fitted onto the protruding stoma. It was also believed to be the best method for preserving renal function and avoiding electrolyte derangements. Only on long term follow-up did it become apparent that stomal stenosis was so common that revisions were required in at least 40% of the children. Some needed two or more revisions. In addition intrinsic strictures developed in the loop after 10-15 years and reflux, along with infection, resulted in chronic pyelonephritis which not only retarded renal growth and development, but it affected renal function.[56,57]

Colon Conduit

In 1975, Hendren[58] proposed the isolated sigmoid colon conduit as a method for temporary external diversion since it is less common for the colonic stoma to stenose and a nonrefluxing ureterocolic anastomosis can be made. In patients with bladder exstrophy who could not be reconstructed he later anastomosed the stomal end of the conduit into the sigmoid colon after confirming that the ureterointestinal anastomosis was unobstructed and nonrefluxing and the child had gained the ability to control liquid stool. In animal experiments, Richie *et al*[59] demonstrated a much reduced incidence of histologically proven pyelonephritis with a nonrefluxing colon conduit as compared to a freely refluxing ileal conduit. In addition, while the colon conduit is exteriorized, hyperchloremic metabolic acidosis is uncommon since urine does not remain in contact with colonic mucosa for any length of time. For the rare case that requires permanent urinary diversion we prefer to use the sigmoid or the ileocecal conduit.

Ileocecal Conduit

The ileocecal conduit also has a low incidence of stomal stenosis, its blood supply is constant and the vessels can be mobilized easily along with a long mesentery. Even in obese patients with a thick mesentery the vessels

are easily identified by palpation. In addition the cecum withstands voiding pressures well in case subsequent undiversion is planned. Originally the antireflux mechanism in the ileocecal conduit was made by plicating the ileocecal valve to increase its competence and (after removing the appendix) by wrapping the cecum around the terminal ileum as is done in a gastric fundoplication. A particular advantage of this conduit is that large ureters can easily be anastomosed to the end of the ileum without having to make a nonrefluxing anastomosis. Unfortunately, long term follow-up has demonstrated that eventually the above mentioned anti-reflux mechanism tends to fail unless the ileocecal valve is intussuscepted into the cecum.[60]

Continent Diversion

The next logical progression in permanent diversions was the creation of a continent catheterizable pouch. The first such attempt was made by Verhoogen.[61] The principle of using the appendix as a cutaneous stoma and making the cecum into a reservoir was first successfully utilized by Makkas.[62] In 1950 Gilchrist *et al*[63] modified this procedure by tunneling the ureters into the cecum to create nonrefluxing anastomoses.

Principles

Continent diversions consist of a reservoir and a catheterizable continent stoma.

Reservoir

It is essential to make a low pressure, high capacity reservoir, similar to the normal bladder. This has been achieved by detubularizing the bowel. The segment of bowel is opened along its anti-mesenteric border and reconfigured into a hollow sphere. According to Laplace's law, such a geometric configuration produces the lowest pressure within the reservoir for a given surface area. Detubularization may further lower reservoir pressures by rendering motor activity in that segment of intestine to become uncoordinated. The latter, however, is believed to be a self-limited phenomenon since the bowel segment regains its peristaltic integrity within three months.[48]

Continence mechanism

The continent ileostomy was popularized by Kock.[64] There are four general categories of continence mechanisms:

a) Intussuscepted nipple

b) Plicated, imbricated or intussuscepted ileocecal valve

c) Mitrofanoff principle (appendix)

d) Reverse intussusception (Benchekroun nipple)

Fundamentally, however, they are either flap valves or nipple valves. The nipple functions because pressure within the reservoir acts on it from the sides and compresses it shut. Unfortunately pressure within the reservoir also displaces the nippe's lateral walls outwards at the base resulting in its gradual effacement with eventual failure of the continence mechanism. In flap valves, on the other hand, the conduit runs in a submucosal tunnel hence it is supported in its length by the wall of the reservoir, and pressure within the reservoir compresses the lateral walls of the conduit and pinches it shut. Furthermore, since the stoma is placed at an angle, it does not become effaced.

Intussuscepted Nipple

In Kock's[64] original operation a GIA stapler was utilized to provide four quadrant fixation. He made no windows in the mesentery. This type of nipple was prone not only to prolapse but it eventually became incontinent once the nipple shortened to less than 3 cm. The shortening is believed to be due to separation of the walls of the pouch and chronic ischemia.[65] In an effort to reduce these complications, Skinner *et al*[66] modified Kock's operation by making an 8 cm window in the mesentery and by using a thoracoabdominal (TA-55) stapler to stabilize the nipple. He also removed the distal five rows of staples to reduce the chances of calculus formation. This modification did reduce the incidence of nipple failure but urinary leak from fistulae and stone formation on exposed staples was reported. Later, Skinner further modified the technique by including fixation of the nipple to the wall of the pouch with a fourth row of staples, the use of Dexon® mesh instead of Marlex®, and the use of a Marlex® strut to prevent failure of the efferent nipple and development of parastomal hernias.[67] In the latest publication by Skinner *et al*[68] only three rows of staples were used. The posterior row incorporated the

wall of the pouch while the two anterior rows stapled only the nipple. Another modification of the intussuscepted nipple was introduced by King *et al*.[69] They approximated the muscular coat of the pouch to the seromuscular layer of the cut edge of the outer wall of the nipple and the seromuscular layer of the inner wall of the nipple. The mucosa of the nipple was also sutured to the mucosa of the wall of the pouch.

Plicated, Imbricated or Intussuscepted Ileocecal Valve

Gilchrist *et al*[63] relied on the inherent resistance of the ileocecal valve along with peristaltic action of the terminal ileum and oblique tunneling of the ileal segment through the abdominal wall to achieve continence. Although they had excellent results on short and long term follow-up[70,71] other surgeons were unable to achieve the same degree of continence. In an attempt to increase the native resistance of the ileocecal valve various techniques have been devised. These have been discussed earlier in the section on the ileocecal conduit. Mansson[72] reported good results when he made an ileoileal rather than ileocecal intussusception.

Mitrofanoff principle (appendix)

In 1980, Paul Mitrofanoff,[73] perfected the technique of making a continent stoma out of the appendix and he popularized its use. However, it should be remembered that Verhoogen[61] and Makkas[62] had previously used the appendix for this very purpose although they could only boast a 50% continence rate. The ideal mechanism is one which responds to increasing pouch pressures by dynamically raising the resistance within the conduit. This principle has proven to be consistently successful in ureteroneocystostomies and tunneled uretero-enterostomies. It has been demonstrated that intraluminal pressures within the conduit located in a submucosal tunnel are two to three times higher than that within the reservoir at full capacity. The appendix is typically used for the procedure. If necessary, added length can be obtained by tubularizing the cecal cuff or a longitudinal segment of the antimesenteric cecum. The stoma should be located at a site that is convenient for the patient to catheterize. It may be placed in the right lower quadrant of the abdomen or in the umbilicus as described by Duckett and Snyder.[74] Not only is an umbilical stoma hidden from view, but it is the one location where subcutaneous fat is not deposited regardless of how obese the patient may be. If the appendix is used for the catheterizable channel and it is placed at the umbilicus, it is important to carry any midline abdominal incisions to the left of the umbilicus to keep the vascular pedicle of the appendix from crossing the incision.[75] The Mitrofanoff procedure has now been expanded into the Mitrofanoff principle. It is the term used to describe the process of making a continent catheterizable stoma for any reservoir by implanting a tube with a narrow lumen into a submucosal tunnel in the reservoir. The appendix still remains the organ of choice but not only must it be present in a given patient, it has to be long enough and of sufficient caliber. The appendix has also been rendered continent by invagination rather than amputation and reimplantation.[76] Finally, the appendix has been amputated at its base and reimplanted in the ileocecal valve.[77] When the appendix has been unsuitable other vascularized structures have been used. These include tapered bowel,[74,78] transversely tubularized bowel,[79] fallopian tube,[80] ureter,[74] stomach,[81,82,83] cecum,[84] defunctionalized colon,[85] and even preputial skin.[86] In patients with a large bladder capacity a nonrefluxing tube can also be made from the detrusor muscle itself.[87] If a detrusor tube is used bladder mucosa exposed to air tends to dry and develop exuberant growth with eventual stenosis, just as has been the experience with bladder mucosal grafts in hypospadias repairs. We avoid the problem by constructing a short skin tube at the stoma, which is anastomosed to the detrusor tube below skin level.

The appendix permits development of the longest functional profile length as demonstrated on pressure profilometric studies of conduits.[88] The ureter is not as efficient and it also seems to be associated with greater discomfort on catheterization[74] and higher complication rates.[89] When segments other than the appendix are used it is imperative that the functional Mitrofanoff profile be at least 2.0 cm. in length.[88] This issue should be given particular consideration when ureter is implanted into bowel. Another important consideration in using ureter for the conduit is preservation of its distal blood supply when reimplanting it. An extravesical approach, such as the Lich-Gregoir technique[90] should be chosen.

Reverse Intussusception (Benchekroun Nipple)

The fourth method is perhaps the most technically challenging and employs a hydraulic valve, the leaves of which close as the pouch fills. This mechanism is a form of reversed intussusception, known as the Benchekroun nipple.[91] The surgical technique involves isolating a segment of small bowel. A reverse intussusception is made by juxtaposing the mucosal surfaces of the small bowel. This valve allows urine to flow freely between the leaves of the bowel mucosa, and as the pouch fills, hydraulic pressure compresses these leaves. This valve is uncommonly used because it is difficult to construct.

Classification of Pouches

They could be external or internal.

External

These pouches have an external catheterizable stoma.

Ileal Pouch-Kock

The Kock pouch is constructed from a long segment of ileum. The part that will form the reservoir is folded into a U configuration and an intussusception valve is created at each end. One valve prevents leak of urine from the surface stoma while the other prevents reflux into the ureters and kidneys. Unfortunately the procedure has a high failure rate. Kock himself reported an 80% failure rate in his original series and today it appears to be about 15% even in the hands of experienced surgeons.[65] The reasons why nipple valves fail have already been discussed. In addition difficulty in catheterization, parastomal hernias, stomal complications and electrolyte imbalances have been reported. All told, the postoperative complication rate may be as high as 30%. Most of these patients also have asymptomatic bacilluria, however, this does not lead to pyelonephritis unless there is reflux into the ureters.

Ileo-Ceco- Right Colon Pouches- Mainz, Penn and Indiana

All of these pouches are modifications of the Kock pouch.

The Mainz pouch was described in 1985.[92] In 1988 they added an ileoileal intussuscepted nipple of the type described by Mansson[72] to further improve continence. The Mainz pouch has a lower reoperation rate than the Kock pouch, however, its reservoir pressures are higher than in pouches made from small bowel alone.[65]

The Penn pouch was described by Duckett and Snyder.[93] It is relatively easy to form, has a reliable continence mechanism and a minimum of small bowel is used. Duckett and Snyder[93] amputated the appendix at its base, reversed it and reimplanted it into a submucosal tunnel in the cecum. They also described the umbilicus as the ideal site for the stoma. Reidmiller *et al*,[94] on the other hand left the appendix attached and embedded its base in a 4-5 cm submucosal tunnel. In our opinion, a combination of the Penn pouch with use of the appendix in situ appears to be the simplest and most reliable method of continent diversion.[95] In case the appendix is too short, a segment of cecum, in continuity with the base of the appendix is developed and tubularized in the cephalad direction. This tubularized segment is buried into a seromuscular tunnel in the medial wall of the colon and serves as the initial part of the conduit.[96]

The Indiana pouch[97] is a modification of the Gilchrist reservoir.[63] To improve continence and to make catheterization easier they tapered the ileal segment and plicated it to the ileocecal valve before bringing it out as the stoma.

Colon Pouch

The detubularized sigmoid colon has also been used for, both, continent diversion and creation of a neobladder.[98,99,100,101,102] The ureters are implanted in submucosal tunnels and a catheterizable stoma is made by implanting the appendix or a segment of ileum into a submucosal tunnel in the pouch.

Gastric Pouch

Gastric pouches can also be used for continent diversion. Since the stomach is generally used for bladder augmentation and occasionally for orthotopic diversion it will be discussed later.

Internal

Urine could also be diverted internally either to an ectopic location such as the colon or into a reservoir that replaces the native bladder.

Ectopic urinary diversion

The prime example of such diversion is the ureterosigmoidostomy of the early 20th century in which the competent anal sphincter was the continence mechanism.[103] Originally employed in pediatric patients with bladder exstrophy and in adults after cystectomy for bladder cancer, there are still occasional indications for this type of diversion. Many of the long-term complications of ureterosigmoidostomy are common to other conduits,

including metabolic acidosis, pyelonephritis, and upper tract deterioration. One problem specific to such a diversion was the inability of some children to control liquid stools that ensued due to the admixture of urine with stool. This was more evident in children who had the procedure performed before they achieved fecal continence. Delaying the procedure until they had achieved continence reduced the incidence of inadvertent leaks from the rectum. These children can also be evaluated preoperatively by administering a tap water enema, which the child is asked to hold for 2-3 hours. If he can keep the enema fluid from leaking out he should be able to control liquid feces. The most bothersome complication was their increased risk for colonic neoplasms. The majority of the tumors are adenocarcinomas, with a mean latency period of 25 years. Thus, long-term follow up and periodic colonoscopy are mandatory. Nevertheless, there are many patients today who have done well with ureterosigmoidostomies, and the technique remains a viable alternative in an appropriate patient.

An ileo-anal reservoir can also be used under specific circumstances. In this procedure, a reservoir is fashioned from ileum and it is attached to the rectum.[104] This procedure has never been popular.

Orthotopic Diversion or Substitution Cystoplasty

The first reported efforts were those of Camey *et al* in 1958.[105] Camey's original technique entailed the use of a U-shaped loop of ileum. The mid-portion of this loop was anastomosed to the urethra. The ureters were anastomosed to either end in an anti-refluxing fashion. The patient's native continence mechanism was retained and the reservoir was emptied by a Valsalva maneuver every two to three hours since Camey's operation predated the institution of CIC.

All orthotopic diversions are associated with nocturnal enuresis to some degree for two main reasons. First the spinal reflex arc, that recruits the external sphincter, is lost during cystectomy. Thus, the patient retains the conscious desire to void, but it is not followed by an involuntary increase in sphincter resting pressure. Secondly, there is a significant shift in urinary metabolites that results in increased urine production after construction of a neobladder.[91]

Nocturnal enuresis is often managed by having the patient awaken two to three times a night to void. Other treatment options in males include a condom catheter or a penile incontinence clamp. Finally, various sphincter operations may be considered. Interestingly, females who undergo this type of procedure often experience hyper continence and may require temporary CIC. Female patients should therefore be followed closely and reassured that this phenomenon usually plateaus after two years.

There are many variations on the original theme of orthotopic diversion. The Camey II operation[106] involves detubularizing a length of ileum to dampen peristalsis and prevent high pressure waves that can cause wetting or back pressure. The pouch of the ileal neobladder described in Ulm, Germany,[107] has a relatively large capacity and an excellent outcome with regard to urinary continence.[91]

In general all the continent diversion techniques described earlier can be modified so as to connect the end that formed the external stoma to the native urethra or to even form a neourethra. In situations where it appears that urethral continence cannot be achieved, the reservoir can be placed in the location previously occupied by the native bladder and a catheterizable conduit could be brought out on the abdominal wall.

The hemi-Kock operation was designed by Ghoneim and associates.[108] Similar to its catheterizable counterpart, this procedure requires construction of a nipple valve to prevent reflux. However, a shorter segment of bowel is needed as only one intussuscepted valve is required. Continence is maintained by the patient's own sphincter. The low pressure bladder substitute described by Studer and coworkers[109] is a variation of the hemi-Kock procedure and it does not require a nipple valve. Although the uretero ileal implants reflux freely, isoperistaltic integrity of the proximal limb (ileal ureter) serves to dampen the effects of a full reservoir. This innovative approach avoids the formation of an antireflux valve with its inherent complications.

In addition to meticulous surgical technique, the overall success of catheterizable pouches depends on careful patient selection. These diversions should not be offered to patients with limited hand-eye coordination, or the inability to comply with the catheterizing process for any reason. Also, continent diversion is not an appropriate choice for patients with a creatinine clearance of less than 60 mL/min[91] According to Benson[91] “The patient should be advised that, all other considerations being equal, continent diversion is associated with a longer hospital stay, a higher complication rate, and a greater potential for reoperative surgery.”

Complications of Urinary Diversion

Complications can be categorized as general complications of major surgery, those due to use of gastrointestinal (GI) segments in the urinary tract and those related to removal of the gastrointestinal segment from the G.I. tract.

General complications

The incorporation of bowel in the urinary tract carries with it the inherent risks of intestinal anastomoses. There is no significant difference in the incidence of fistula formation, whether the anastomosis is stapled or hand-sewn. Sepsis is common if a fistula develops and is associated with a mortality rate of approximately 2%.[48] Bowel obstruction due to adhesions is also relatively common, the incidence being slightly less if large bowel is used. Small bowel obstruction occurs in about 2.5%-3.5% of patients. The largest series,[110] of bladder reconstructions using bowel consisted of 231 patients. Eight of their patients (3.5%) developed bowel obstructions. Hendren[111] reported 2 cases of bowel obstruction out of 68 bladder augmentations for undiversion (3%). One should keep in mind that tumor recurrence may also present as a bowel obstruction in patients originally operated on for a.malignancy. Hemorrhage and pseudo-obstruction are also well recognized complications of intestinal anastomoses.

There are a host of complications associated with the creation of a stoma, and stoma related problems are typically the most troublesome in the distant postoperative period. There is a 20%-24% incidence of stomal stenosis in ileal conduits, and a 10%-20% incidence in colon conduits.[48] Stomal retraction, prolapse, and parastomal hernia are potential problems that may necessitate revision of the stoma. Chronic dermatitis or necrosis of the stoma site with associated bleeding, though not fatal, can be bothersome.

Use of GI Segment in the Urinary Tract

The use of isolated intestinal segments for urinary diversion is classically associated with well described electrolyte imbalances. The benefits and disadvantages of various GI segments when used in the urinary tract are summarized in Table 1. The jejunum is generally no longer used as its very highly absorptive surface causes severe metabolite shifts. Historically, jejunal conduits caused a hyperkalemic, hyponatremic metabolic acidosis. Ileal segments result in a hyperchloremic metabolic acidosis which is thought to be the result of active exchange of ammonium chloride for carbonic acid. This mechanism involves a chloride pump that is dependent on cyclic AMP (cAMP). The metabolic acidosis may therefore be treated with agents that inhibit cAMP, such as chlorpromazine or nicotinic acid. More commonly, sodium bicarbonate is tried initially. This complication is seen in about 15%-20% of patients, but not all of them require treatment.

TABLE 1. Comparison of G.I. Segments In Bladder Augmentation

Intestinal Segment	Advantages	Disadvantages
Ileum	Most popular and technically simplest; most compliant, least contractile	Reflux into ureters. Hyperchloremic metabolic acidosis from long term exposure.
Ileocecal segment	Large area, can be converted to conduit if it fails; anti-reflux via intussusception of valve	Severe diarrhea in 10-15% of patients with myelodysplasia. Valve tends to fail over time.
	Thick submucosa for reimplantation, large lumen, easy to mobilize sigmoid colon.	Greatest mucus production and most contractile; ?Long-term risk of malignancy greater; hyperchloremic, hypokalemic metabolic acidosis
Stomach	Thick submucosa; useful in renal insufficiency; lowest incidence of calculi and UTIs; low mucous production	Hematuria-dysuria syndrome in 20-25%; hypochloremic metabolic alkalosis with long-term exposure

Colon conduits are generally associated with similar electrolyte abnormalities as the ileum. Hypokalemia and total body potassium depletion may also occur but it is usually mild in nature. Unfortunately, the sigmoid colon is more prone to diverticulosis and malignancy, and may not be suitable for long term urinary diversion.

Gastric reservoirs have unique properties resulting in a distinct metabolic profile. Gastric mucosa acidifies the urine and generates a net excretion of chloride ion. Patients may present with hypochloremic metabolic alkalosis that is usually more pronounced in patients with advanced renal insufficiency and is much more difficult to treat. The combined use of stomach and small and/or large bowel has been used to balance the negative side effects of each segment.[112]

Mechanical complications are also described for isolated intestinal segments. The conduit may develop a stricture due to lymphoid depletion of the intestine and the accompanying inflammatory response from chronic exposure to urine. Conversely, the segment may become elongated, particularly if there is distal obstruction.

It has been noted that even after detubularization of right colon, reservoir pressures can be as high as 60 to 110 cm of water.[113] In one study, 42% of patients with detubularized ileum and 60% of patients with detubularized right colon had contractions greater than 15 cm of water although clinically significant contractions, in which pressures rose higher than 40 cm of water at volumes of less than 200 ml, were only found in 10% of detubularized right colon and in none of the detubularized ileal segments.[114] This study suggests that ileum is superior to right colon when large volumes of urine are involved. Cecal reservoirs have ten times greater motor activity and higher basal pressures than ileum[115] It has been suggested that placing a second patch of bowel on the reconfigured bowel reservoir will eliminate return of contractile activity and it will also increase the volume of the reservoir.

Removal of GI Segment from Continuity

Removal of small bowel and the ileocecal valve from the GI tract also cause problems. Particularly affected are patients with myelodysplasia who maintain fecal continence by being constipated. Removal of the ileocecal segment in these individuals can result in intractable diarrhea. Other complications caused by removing terminal ileum from the G.I. tract include diarrhea and vitamin B12 deficiency, but they are rare and tend to occur only if large segments of terminal ileum are removed.[116]

Right and sigmoid colon procedures offer some theoretical advantages over small intestine. The enterohepatic circulation is preserved if ileum is left in situ. In addition, a sigmoid pouch can be brought down to the membranous urethra with relative ease, and removing colon from the G.I. tract has little impact on the nutritional status or bowel habits of the patient. Colon conduits produce fewer nutritional disturbances, provided the ileocecal valve is not violated.

Removing a segment of the stomach generally does not cause any long-term problems although early satiety is noted in all these children in the immediate post-operative period. Vitamin B12 deficiency has been reported with use of an extremely large segment of stomach.[116]

Urinary Undiversion

Some of the sequelae, of long-term urinary diversion such as, metabolic acidosis, hydronephrosis, chronic bacilluria, low-grade sepsis and gradual upper tract deterioration are more pronounced in children with severe obstructive uropathy or lower tract dysfunction.[117] Thus, although urinary diversion became the accepted form of surgical management for these children in the 1950s and 1960s, it left them at risk for numerous complications. In addition, the need for an external appliance throughout childhood and adolescence had a serious impact on their quality of life and self-image.

Undiversion is always a complex, challenging and time consuming procedure which requires complete and thoughtful evaluation of each patient as an individual followed by use of the best option available in a given situation. The surgeon who wishes to undivert a patient must not only be technically adept with a wide armamentarium at his disposal, but he/she must be able to change plans during the course of the procedure, if necessitated by the operative findings.

History

Dr.W.Hardy Hendren III coined the term "undiversion" and pioneered refunctionalization of the previously diverted urinary tract.[118] Having been dissatisfied with long term results of urinary diversion, he had previously

proposed that primary reconstruction, rather than diversion, was the better option in a series of 63 patients with decompensated megaureters.[119] His excellent results with primary reconstruction led him to carry out anatomic and extra-anatomic reconstructions in children whose urinary tracts that had been diverted for varying periods of time. Initially undiversion was limited to children diverted for non-neurogenic urinary tract pathology (*i.e.* PUV, prune belly syndrome and VUR) and whose renal function was still preserved. After Lapides[33] demonstrated the safety of nonsterile or clean intermittent catheterization (CIC), the indications for undiversion were expanded to include children with neurogenic bladders secondary to myelodysplasia and even to patients awaiting renal transplantation.

Initially, all undiversions were carried out with the native bladder in place and with the expectation that the child would either void or catheterize per urethra. Today, continent diversion and bladder substitution are also utilized to rid the child of an external appliance and to protect the upper urinary tract.

Indications

The most common indication for undiversion is progressive hydronephrosis and upper tract deterioration even though this may have been the reason why the urinary tract was diverted in the first place. Renal damage is usually due to chronic pyelonephritis caused by reflux of infected urine from the conduit to the upper tracts. In addition, stenosis at the ureteral anastomosis, within the conduit or at the stoma can lead to chronic obstruction and hydronephrosis.

Undiversion should also be considered if stomal problems (prolapse, hernia) contribute to a poor quality of life for these children.[120] Self-esteem and confidence are low when one is relegated to wearing an external appliance for much of childhood and many patients are subjected to derogatory remarks from other children, particularly if they smell of urine. Cumming[121] reported that single patients were more likely to pursue undiversion to improve self-image and sexuality.

Menon[122] recommended that undiversion be offered to all patients with neurologically intact bladders who were diverted previously for the above reasons and even some with neurogenic bladders. Mundy,[123] on the other hand, found there was "rarely a reason to undivert a wheelchair bound patient with a functional permanent diversion"; this was, however, prior to the era of the appendicovesicostomy and its benefits.

In early reports renal failure was thought to be a contraindication for undiversion. Richie and Sacks[124] suggested that only children with a creatinine clearance greater than 40 mL/min or stable renal functions were candidates for undiversion, and that inappropriate patient selection (*i.e.*, poor renal function) could result in problems after undiversion. On the other hand Gonzalez[125] proposed that even children with end-stage renal disease and those awaiting renal transplantation should be undiverted. He was of the opinion that re-establishing the urinary tract prior to transplantation increases both, bladder capacity and continence (especially if an adult kidney with a higher urine output is grafted in a child). In addition, he showed that chronic infection resolved after undiversion. Finally, he stated that early reconstruction allows for adequate healing prior to placing the child on immunosuppression.

Prolonged diversion should not be viewed as a barrier to undiversion. Goldstein[126] reported on seven adult patients who underwent total reconstruction of the urinary tract having been diverted for a mean duration of 14.8 years - one patient had been diverted with a nephrostomy tube in a solitary left kidney for 38 years! Hendren[127] undiverted 177 patients, 32% of whom were diverted for ten years or more, one for 27 years. Such cases demonstrate that undiversion is a viable option even when the urinary tract has been dormant for decades.

A small capacity bladder is no longer considered to be a contraindication to undiversion since many of these bladders can be salvaged by bladder cycling, judicious use of anticholinergics prior to undiversion and bladder augmentation. It is currently felt that, if at all possible, it is best to use either the whole bladder or a part of it for undiversion rather than to make a continent diversion with bowel alone.

Contraindications

One absolute contraindication for undiversion is the noncompliant patient. This subject will be discussed further under patient selection. Undiversion is also to be avoided in patients who are incapable of intermittent catheterization due to inadequate hand-eye coordination. Patients with marked abdominal sensory loss are at greater risk since complications may go undetected for long periods of time. Finally, a patient with severely diminished mental function will not benefit from undiversion, unless he or she has a dedicated caregiver who is willing to address the rigors of caring for the undiverted tract.

Preoperative Evaluation

Patient Selection

Not all patients who are diverted will accept or are candidates for undiversion. Before undiversion can be considered the primary pathology (*e.g,* PUV), that led to diversion in the first place, should either be resolved or addressed definitively. The child must be motivated enough to undergo a lengthy operation and to return for regular postoperative visits. The patient and his parents must also be prepared to cope with potential complications, including reoperations and even re-diversion in the event of failure. Preoperative psychological evaluation is recommended.[127,128] In addition, the patient must understand that he may never be able to void spontaneously or adequately after the reconstruction and must be willing to learn CIC prior to the operation. If the patient is not motivated or unwilling to accept these responsibilities, undiversion must not be offered as an option.[129]

Functional Evaluation

The role of formal urodynamics prior to undiversion is unclear. While some authors feel that all children need preoperative urodynamic studies,[127] others consider it necessary only in those with myelodysplasia.[120,121] Bladder cycling in lieu of formal urodynamics has been proposed as well.[134] The bladder is cycled by intermittently filling the native, diverted bladder with sterile saline or neomycin solution[130] either through a suprapubic cystostomy or during CIC. The contracted, small capacity bladder often enlarges to a size that will support the urine flow after undiversion and will become continent, thus obviating the need for augmentation cystoplasty with bowel segments. Wacksman[131] demonstrated that, in some cases, the bladder will "cycle itself" once urinary tract continuity is restored and it will expand to a reasonable capacity without preoperative cycling. We believe that preoperative bladder cycling is essential in preparing a patient for undiversion. Apart from being a good measure of the patient's motivation and ability to self-catheterize it may decrease the size of the augment or avoid the need for augmentation.

Bladder cycling can also be used to get a reasonable indication of postoperative continence. The child is instructed to fill the bladder until he senses fullness and then to ambulate, if possible. If the child is able to hold the fluid without leaking, it is reasonable to expect continence after urine flow is restored.

Adequacy of sphincter function can be determined by an electromyogram (EMG) during formal urodynamics. A leak point pressure of greater than 40 cm H2O is indicative of adequate outlet resistance and postoperative continence.[128] However, even those with low leak point pressures, can be rendered continent with bladder neck reconstruction.[43,132]

The length of time required to obtain the maximum benefit from bladder cycling is controversial. Firlit[133] noted an increase in bladder capacity after cycling for a mere five days, and Ahmed[134] felt that one week was enough. Obviously, the more contracted the bladder the longer it will have to be cycled. Perlmutter[135] has reported that cycling may be required for up to nine months preoperatively. In our experience a period of 2 - 3 weeks is needed for the patient to achieve its maximum benefit. If cycling does not result in a significant increase in capacity, augmentation cystoplasty must be considered, however, use of a bowel segment should be tempered or even avoided in patients with existing renal insufficiency in light of the metabolic strain that may ensue.[134] Stomach is the best choice if the patient's glomerular filtration rate is below 30mL/min.

Radiographic Evaluation

All patients must have the anatomy and function of the existing diversion evaluated thoroughly. Intravenous pyelography (IVP), when kidney functions permit, and a retrograde contrast study of the conduit (loopogram) to demonstrate reflux and/or obstruction are mandatory. A renal scan is useful if poor function is suspected in one of the renal units and to determine whether a dilated collecting system is truly obstructed or only dysmorphic. A voiding cystourethrogram (VCUG) helps determine the presence of VUR, and bladder outlet obstruction. It can also be used to measure bladder capacity.[136] Careful visualization of the bladder neck and urethra on a VCUG is helpful in predicting whether the patient will need concomitant bladder neck reconstruction. Video urodynamics is useful in patients with myelodysplasia to test urethral competence[137] and also to determine bladder capacity in the presence of dilating VUR. Retrograde ureterography during cystoscopy, especially if there is no VUR, is useful in assessing the length of the ureteral stumps and their adequacy for primary anastomosis of one or both ureters.

Lower Tract Evaluation

Much of the evaluation of the native bladder and urethra has been addressed in the section on radiographic evaluation. Cystoscopy is essential in the preoperative assessment of the diverted bladder. Other than the usual small, contracted bladder, one can also expect to encounter pale mucosa that is very friable and bleeds easily.[137] The ureteral orifice(s) should be cannulated to assess the length of the submucosal tunnel and the need for possible simultaneous ureteroneocystostomy. The configuration and location on the trigone of the orifice(s) should also be identified. Hendren[138] stated that if one or both have a "golf hole" appearance, it was suggestive of high-grade reflux and reimplantation would be required.

Outlet resistance is an important component of a proper urodynamic study and a thorough evaluation of infravesical functional status is mandatory. High voiding pressures within the bladder can be expected if the resistance is too high, as in detrusor-sphincter dyssynergia.[136] In such cases, alpha-blockers or muscle relaxants to relax the smooth muscle at the bladder neck along with CIC will be needed postoperatively to aid in bladder emptying. Botox injections are also a useful modality and in the most severe cases, sphincterotomy to reduce outlet resistance may be indicated if nonoperative measures fail.

In contrast, inadequate resistance is common in myelodysplastic patients, especially at the level of the bladder neck and proximal smooth muscle sphincter. Sphincter EMG, along with a urethral pressure profile, is helpful in assessing function of the striated external sphincter, which may be the only means of maintaining continence in these patients.

Upper Tract Evaluation

As part of the preparation for undiversion, it is essential to ensure that existing tissues are used in the safest, most efficient manner to restore the urinary tract. Good ureteric function is critical to the success of any undiversion.[139] Any dysfunction that is not addressed could result in obstruction or reflux, both of which could lead to silent upper tract deterioration, often the reason that undiversion was pursued in the first place. Pressure flow studies of the distal ureters can be performed with small balloon catheters inserted into the ureteral stumps to rule out distal obstruction.[132] It is also essential to evaluate the proximal ureter and UPJ. These structures are usually evaluated by ultrasonography or renal scans.

Laboratory Evaluation

Even though renal insufficiency is not a contraindication to urinary undiversion, it is imperative that renal function be evaluated preoperatively to permit use of an appropriate G.I. segment. Serum electrolytes, blood urea nitrogen, creatinine and creatinine clearance (using a 24 hour urine sample) should be measured as a baseline study and followed closely in the postoperative period. Collaboration with a nephrologist is helpful in managing renal functions in the long-term. Normal liver functions preoperatively must be documented, as patients with hepatic insufficiency are at risk of ammoniagenic coma if bowel is used for undiversion.[116]

Surgical Principles

There are some basic, universal tenets that apply to any undiversion. In general these are long, tedious and complex cases that may last for 8 to 12 hours or more. A full mechanical and antibiotic bowel preparation is always essential since the final choice of bowel segment to be used is only possible on the operating table. Broad-spectrum antibiotics to cover anaerobes and intestinal aerobic bacteria are given pre and postoperatively. Postoperative nasogastric decompression is always required even if intestinal resection and anastomosis is not performed since prolonged exposure and handling of bowel invariably results in an ileus. Abdominal distention from ileus could compromise the augment by compressing its pedicle. Central venous access is secured not only for intraoperative fluid infusion but for postoperative nutritional support. The incision should be generous and often extends from the xiphoid to the pubis. Inadequate exposure and struggling for exposure are not only tiring but result in substandard surgery.

In the urinary tract, it is of paramount importance that the anastomosis be water-tight and free of tension, to prevent postoperative urinary leakage.[131] In addition, a postoperative drain is left adjacent to but not in contact with any anastomoses. Adequate mobilization of tissues is helpful in achieving a tension-free repair. While mobilizing the ureters their blood supply must be carefully preserved to prevent ischemic strictures. This is best attained by a wide dissection and by sweeping retroperitoneal tissues towards the ureters. It is generally accepted that reflux into the upper urinary tract should be prevented.[124] A few authors[122] believe that formation of an anti-reflux mechanism is not essential in a compliant, low-pressure bladder, however, a compliant, low pressure bladder is a rare entity in the diverted patient, especially one with myelodysplasia. Whether ureter or a bowel segment is reimplanted, the submucosal tunnel should have a diameter-to-tunnel length ratio of 1:5 to

provide an adequate flap valve to prevent reflux. It is also best if the ureter is reimplanted into a fixed part of the native bladder (*i.e*, the trigone).

Augmentation Cystoplasty

Most authors agree that the native bladder should be part of the continent reservoir, if at all possible.[123,127] Augmentation cystoplasty using a segment of bowel was not utilized in early undiversion cases, especially in those with compromised renal function.[120] Many patients were able to attain normal or near normal capacity and urinary continence with bladder cycling alone. However, in time, it became clear that some patients remained incontinent as they had bladders of low capacity and poor compliance. This was especially true in patients who had a high urine output, such as those following renal transplantation[43] or with poor concentrating ability of the native kidneys. Since patients with obstructive uropathy may also have a concentrating defect (Type IV renal tubular acidosis) it is imperative to know the baseline urine output before attempting augmentation so as to decide on an appropriate reservoir. Measuring the urine output for a few days helps determine the average urine output in ml/kg/hr. This knowledge would also be valuable in determining the interval between catheterizations after augmentation.

If the patient has a continent bladder neck and an intact trigone the augment is placed on top of the native bladder. In such instances the detrusor must be made incompetent or it will contract and result in an hour-glass shaped reservoir which will develop high intraluminal pressures. Detrusor contractions can be eliminated by either excising the dome of the bladder so that the trigone and lower part of the bladder attain a funnel like shape or by bivalving the bladder. In poorly compliant bladders bivalving increases the surface of the anastomotic line and decreases the chance of a stricture developing and resulting in an hour-glass deformity. Bivalving also allows for maximal bowel to bladder contact and decreases the anastomotic length of the bowel to bowel segment thus approximating a sphere rather than a "clamshell".

In incontinent patients, the trigone and lower part of the bladder are used to develop a continence mechanism and the augment is sutured to the small cuff of retained bladder above it. Of course, when it is decided to replace the entire bladder, the cystectomy is carried out at the junction of the bladder with the urethra and the bowel segment, with its own continence mechanism, is anastomosed to the native urethra.

In all bladder augmentations, a urethral catheter and a suprapubic tube are left for drainage. If the ureters are reimplanted or at risk of edema developing double J stents may be placed. If possible, all externalized tubes are brought out through the wall of the bladder segment, not the intestine. In addition the area is drained and the peritoneum is mobilized and made to cover the augment so as to isolate the reservoir extraperitoneally.

Various segments from the gastrointestinal tract can be used, as is the case for continent diversion.[140] The advantages and disadvantages of each segment mentioned previously are summarized in Table 1.

Ileocystoplasty

This is technically the simplest of the various options. In Camey's[105] original operation the ileum was configured into a U-shape and the urethra was attached to the apex of the U while the ureters were anastomosed to each end. This technique does not detubularize the bowel. The technique currently in use involves isolation of a 15-20 cm segment of ileum, usually approximately 15 cm proximal to the ileocecal valve so as not to compromise vascularity of the terminal ileum, the ileocecal valve or the cecum. It also has the benefit of leaving the absorptive terminal ileum and the ileocecal valve in continuity, which is of great importance in maintaining fecal continence, particularly in patients with myelodysplasia. After restoring intestinal continuity the isolated segment is opened along its antimesenteric border and reconfigured into a U-shape and the adjacent walls are sutured. The patch is then folded over and the lateral edges are sutured to form an inverted cup, which is then sewn to the native bladder. If the bladder is to be substituted a Kock or hemi-Kock pouch can be used with one end attached to the urethra.

Colocystoplasty

Due to its location close to the bladder the sigmoid colon is most often used for colocystoplasty. The colon segment can be detubularized, as described for ileocystoplasty, prior to suturing it to the bladder. It can also be opened on its antimesenteric border, after suturing the ends closed, and to be placed transversely on the bivalved bladder. Since colon segments produce electrolyte changes and large amounts of mucous, attempts have been made to de-epithelialize the colon segment by mechanical or chemical means prior to augmentation. This permits growth of urothelium into the augment[141,142]

Cecocystoplasty

The cecum can be used as such or after detubularization. The ureters may be reimplanted into the cecum or into a short segment of attached terminal ileum with the ileocecal valve constituting the antireflux mechanism. This segment is never used in myelodysplastic patients but it can be used in patients who were diverted for PUV, exstrophy of the bladder, trauma or tumors.

Gastrocystoplasty

The stomach is an excellent organ for bladder augmentation or substitution in children since it is not usually affected by intrinsic disease nor does its removal from the G.I. tract lead to any long-term sequelae. In addition it is generally out of the radiation field in patients with malignancies.

The stomach is naturally suited for storage and it tends to expand slowly over time. Its musculature produces coordinated contractions, which may contribute to effective elimination of urine from the augmented bladder. Gastric mucosa tends to resemble urothelium most closely since it is not absorptive for most contents, unlike intestinal mucosa, and it is a barrier against organisms. The natural tendency of gastric mucosa to secrete acid helps control the metabolic acidosis that occurs in renal failure. This is a definite advantage over intestinal mucosa, which tends to worsen the acidosis. Gastric mucous is less cellular and viscous than intestinal mucous. Thus catheters do not clog and it may be the reason why stones do not form in gastric reservoirs.[75]

The gastric segment is by no means a panacea. It produces its own unique set of problems, such as hematuria-dysuria syndrome (HDS) and metabolic acidosis, and there is some concern about the development of malignancy. These issues will be discussed later.

The gastric patch may be based on either gastroepiploic artery, but the right gastroepiploic vessels are generally more substantial and it is easier to bring the pedicle down without tension. The appendix can be implanted into the gastric patch for the catheterizable channel or a tube can be made from the central part of the anterior surface of the patch. This tube is made continent by invaginating its base into the lumen of the augmented bladder, and closing the bladder wall around it to create a nipple.

Bagli and Mitchell[75] have described variations of the gastric wedge that can be used to make a catheterizable tube to attach to continent reservoirs made from bowel. They have also made a catheterizable channel from the distal end of the patch and modified the patch so that an artificial sphincter may be placed.

Ureterocystoplasty

Since this technique is relatively new,[143,144] long term results are not available. However, functional storage capacity increases tremendously with this procedure and since urothelium is in contact with urine, metabolic complications associated with G.I. segments are avoided. Initially it was carried out by longitudinally opening a dilated ureter that drained a nonfunctional moiety. Subsequently variations have been described. In one, a dilated ureter from a duplicated system was used.[145] If both poles are functional, a pelviureterostomy or ureteroureterostomy in the upper tract gives a route for egress of urine from the segment from which the ureter is harvested. We have also carried out a ureterocystostomy by using the distal ureter from a functional kidney and draining that kidney into the contralateral side with a TUU. Another modification described utilizes an 8-10 cm segment of lower ureter which is not opened but its upper end is splayed open and anastomosed to the dome of the bladder as if it were the handle of a tea-pot. We have no experience with this method but it should not only increase the bladder capacity significantly but stasis should not occur since both ends drain into the bladder.[146] Ikeguchi *et al*[147] have created a segmental megaureter in a pig model, by using a tissue expander within the ureter. They then successfully used this segment for ureterocystoplasty. It is possible that nondilated ureters may be used in the future for cystoplasty since the ureter has a natural tendency to stretch under pressure. Two special concerns with ureterocystoplasty are that the bladder must be opened in the proper area to avoid development of a "Mickey Mouse" appearance of the bladder. Secondly the ureterocystoplasty obscures the site of reimplantation of the ureter after transplantation.

Autoaugmentation

In essence autoaugmentation creates a large diverticulum of the bladder, which drains easily and completely.[148] The procedure has numerous advantages. Electrolyte imbalances are avoided since urothelium of the bladder is still intact, there are no anastomoses that could leak, and the approach is extraperitoneal. The detrusor muscle is incised in an antero-posterior direction over the dome of the bladder to expose the mucosa, which bulges out. The detrusor on either side is dissected away from the mucosa until a wide mouthed diverticulum is formed. It is

essential to not leave any mucosal perforations. In patients with trabeculated and very hypertrophied bladders perforations do occur during dissection and each one must be meticulously closed with absorbable sutures. Some authors feel that incision of the detrusor (vesicomyotomy) gives as good a result as vesicomyectomy.[149] This procedure has been carried out laparoscopically by both the transperitoneal and extraperitoneal approaches.[150,151] Other authors have buttressed the diverticulum with omentum,[152] or demucosalized stomach[75] and colon.[153] The initial enthusiasm for this procedure has been dampened on follow-up, which has demonstrated reduction in bladder capacity over time.[154] This procedure is now generally used only in patients who have a noncompliant bladder with adequate capacity on preoperative evaluation.[155]

Tissue Engineering and Artificial Bladder

Attempts at augmenting or replacing the bladder with synthetic and biologic materials are not new. In the past synthetic materials like polyvinyl and gelatin sponges,[156,157] Teflon felt[158] and silicone rubber[159] have been used. Biologic materials such as preserved bladder,[160] omentum,[161] peritoneum,[162] dura,[163] fascial free grafts and bovine pericardium[159] have also been tried. These materials have resulted in problems such as lack of union with surrounding tissues, formation of stones, fibrosis, scarring and infection of the graft. At present collagen-based biodegradable materials are being studied for their regenerative potential and functional capabilities.[164] Needless to say, there is no clinically available method of creating an artificial bladder at present.

Incontinence Procedures

The majority of patients who need bladder augmentation are unable to void properly and rely on CIC to empty the bladder. Children diverted for non-neurogenic causes, and some with myelodysplasia, have intact urethral/bladder neck sensation and are unable to tolerate intermittent catheterization. To facilitate catheterization in these children perineal urethrostomy was done, but it proved too cumbersome and uncomfortable.[43] The abdominal continent stoma is a major advance in the management of these children since it is insensate and permits them to catheterize themselves in a sitting position, which is of great value to wheelchair-bound patients. However, the problem of urinary leakage from the native urethra still has to be addressed. One option is to completely close off the bladder neck. While this solves the problem of urinary leakage it also leaves no other route for catheterization. If the patient is unable to catheterize the stoma on occasion and also in noncompliant patients the urethra is a convenient safety valve that prevents over distension and rupture of the bladder. Various procedures have been devised to enhance competence of the bladder neck.

Urethral lengthening

Numerous techniques have been devised to reconstruct the native urethra or to develop a neourethra by using a skin tube, vaginal wall, buttock flap, or even the bladder plate itself under certain special circumstances.[123,127] However, the operation used most often to lengthen the urethra is the Young-Dees Leadbetter procedure. In 1907, Young first increased the functional length of the urethra (1907), subsequently Dees[165] increased mechanical support to the lengthened urethra by incorporating bilateral reduction of the bladder neck, and after that Leadbetter[166] improved reconstruction of the trigonal tube by reimplanting the ureters proximally and wrapping the full thickness of the bladder wall around the tube. Bladder neck reconstruction is often included in the primary reconstruction. It can also be used as a secondary procedure for patients with persistent incontinence.[132]

Flap Valve

Procedures that create a flap valve also lengthen the urethra. In the Kropp procedure a tube is made from the full thickness of the anterior bladder wall and it is tunneled under the mucosa of the posterior bladder wall.[167] This technique is very successful in achieving continence since the tube in the tunnel is compressed as the bladder fills, but difficulty in catheterization is not uncommon. The other technique is that of Pippi Salle *et al*.[168] In this operation anterior and posterior bladder wall flaps are developed. The mucosa of the anterior flap is sutured to the posterior flap to make the neourethra and the muscle of the anterior flap is sutured to the exposed muscle of the posterior bladder wall on either side of the neourethra. Previously mobilized lateral mucosal flaps on the posterior bladder wall are then sutured over it to form the tunnel.

Elevation and Compression of the Urethra

The rectus fascial sling raises the urethra and bladder into a more intra-abdominal position, thus compressing the urethra and transmitting sudden increases in intra-abdominal pressure to the bladder neck.[169,170] Apart from the fact that no artificial materials are used, this procedure has the added advantage that it can be used when other

procedures have failed. There is, however, some question whether the prostatic urethra of post-pubertal boys can be raised and compressed against the anterior abdominal wall.[171]

Mechanical Compression

An artificial urinary sphincter (AUS) produces continence by raising urethral pressure. Best results are obtained if the child has a compliant bladder with a large capacity and is able to void spontaneously. A child who can void by using the Valsalva maneuver or intermittent catheterization may also be considered for the procedure. Its use in children is limited by the need for revision as the child grows or for mechanical failure. The AUS also affects detrusor function which can result in renal failure[172,173,174] and it introduces the risk of infection and erosion into the urethra. Most authors agree that the artificial sphincter should be used only as a last resort if native tissues fail.

Mechanical compression can also be produced by injection of bulking agents in the periurethral tissues. Numerous agents have been used.[175,176,177] Polytetrafluroethylene was the first substance used, however, it caused concern once reports surfaced that it migrated to vital structures.[178] At present we use Dextranomer Hyaluronic Acid (Deflux). It can be introduced transurethrally through a cystoscope, or periurethrally while viewing the urethra with a cystoscope to ensure that the urothelium is not breached by the needle.

Bowel Control

Fecal soiling is a problem in almost all children with myelodysplasia. The basic principle of their management used to consist of producing constipation while ensuring that they evacuated periodically. In the past, dietary changes and constipating drugs were used in conjunction with purgatives and enemas. Accidents were not unusual since purgatives result in uncontrolled evacuation. Enemas produced a more complete and better controlled evacuation but self-administration of enemas is difficult for wheel chair bound children with sensory loss in the perineum, particularly when they become obese. In 1990 Malone[179] described the antegrade continence enema (ACE). By tunneling the appendix into the cecum, a catheterizable, nonrefluxing conduit is made for highly effective antegrade washouts of the colon. We believe this to be a major advance in the management of children with fecal incontinence and constipation and we use it liberally. In patients with a long appendix we leave it attached and tunnel its base into the taenia of the cecum. In other instances it can be amputated with an elongated cecal cuff which is brought out as the stoma while the distal end is implanted into the cecum in a nonrefluxing manner after amputating its tip.

Upper Tract Reconstruction

If at all possible, the native ureter should be used for undiversion. For example, in patients with an existing ileal conduit, primary ureteroureterostomy is the first choice for restoring decompensated ureters.[122] The terminal 1 cm of tissue should be discarded to avoid suturing devitalized tissue and the ends should be spatulated and closed in a single layer to insure a widely patent anastomosis.[130] Indwelling stents assure adequate drainage of urine in the healing phase. It is often difficult to mobilize both ureters for primary repair, especially following an ileal conduit. In these patients the ureter on the side opposite to the conduit is generally short and TUU is a useful adjunct. If the patient requires a concomitant anti-reflux repair, the TUU enables us to carry out one good reimplantation instead of two suboptimal ones.[122]

If one of the ureters is markedly deficient in length, there are several options to bridge the gap. A psoas hitch may elevate that side of the bladder enough to anastomose to the ureter or a tubular bladder flap (Boari flap) can be raised. Boari flaps permit anastomosis of the upper ureter or even the renal pelvis to the bladder. Inferior nephropexy, after mobilizing the kidney on its vascular pedicle, can be used to bridge a gap of up to 3 cm.[134] The conduit itself can be converted into an ileal ureter. Tapering of the segment will maintain structural integrity and decrease mucous production.[131] Waters[180] safely bridged ureteral gaps with ileum in an animal model. None of his six dogs developed azotemia or major metabolic changes. Finally, as a last resort, autotransplantation of a kidney has also been reported, however, it should be avoided in an already abnormal kidney.[181]

Results

Much of the information on urinary undiversion has been presented in the form of case reports to emphasize the complexity and uniqueness of each case. To consider an undiversion successful, the patient must either have improved or stable renal functions, be continent of urine and free of infections, and have an improved or unchanged quality of life. There are several series with long-term follow ups of children and adults following undiversion. It is important to identify the patient population included in each report, since the behavior of the

neurogenic bladder due to myelodysplasia is unpredictable and it can change over time.[182] Also, the definition of "continence" must be scrutinized since varying degrees of dryness are considered acceptable by different authors. Table 2 summarizes the results of various series with respect to continence and renal function.

TABLE 2. Renal Function and Continence After Undiversion

Sexual function

Author (Year)	No.	Improved/Stable Renal Function	Continence	Notes
Hendren (1990)	177	177 (100%)	167 (95%)	44/177 >10 years
Gonzalez (1986)	50	50 (100%)	45 (90%)	3 groups*
Mitchell (1981)	60	N/A	52 (87%)	All augmented
Ahmed (1987)	35	N/A	32 (95%)	32/35 myelo***
Menon (1982)	27	24 (88%)	27 (100%)	8/27 myelo***
Herschorn(1994)	20	20 (100%)	17 (85%)	All myelo***
Frank (1982)	17	17 (100%)	14 (84%)	4/21 neurogenic
Stone (1982)	14	13 (92%)	13 (92%)	Renal failure pts
Cumming (1988)	18	14 (77%)	13 (71%)	3 groups**
Bauer (1980)	14	N/A	10 (71%)	All myelo***
Goldstein (1982)	7	5 (71%)	7 (100%)	3/7 myelo***
Firlit (1980)	20	19 (95%)	N/A	
TOTALS	450	339/350 (97%)	397/430 (92%)	

* included neurogenic bladder, chronic renal failure and neurologically normal bladders
** included neurologically normal bladder, neurogenic bladder and "occult, recovered neuropathy"
*** myelo stands for patients with myelodysplasia.

is another important issue to be considered when evaluating quality of life in adults after undiversion. At present information is limited, but as these children are followed into their adult years, more information on potency, ejaculation and incontinence during intercourse will emerge. This paucity of information is exemplified by a report from Quinif[183] who was able to publish a report on only two undiverted men who had no leakage during intercourse with bladder cycling preoperatively. One patient did well with regard to sexual function after undiversion. However, the other patient, despite full sexual potency after undiversion with ileovesicostomy, faithful intermittent catheterization and maximal pharmacotherapy, had persistent coital incontinence that ultimately led to the patient requesting that he be diverted again. Failure of the proximal sphincter and bladder neck was thought to be the etiology, as the patient also noted retrograde ejaculation coinciding with the onset of urinary incontinence.

Complications

Persistent Incontinence

In the largest study to date,[127] the overall complication rate of urinary undiversion, defined as requiring reoperation for persistent incontinence or reflux, is about 8%, with about 5% (10/177) being related to incontinence alone. Earlier studies often included patients who did not undergo continence procedures initially. It soon became clear that the majority of those undiverted require bladder neck/urethral reconstruction, AUS, or appendico-vesicostomy for persistent leakage. In Ahmed's series,[134] 10 of 35 patients required secondary reconstruction for incontinence or reflux, but he did not consider them complications. In a study of 25 undiverted patients by Mitchell,[43] 6 patients were incontinent and underwent bladder neck plasty to achieve dryness. Nevertheless, 4 of the original 25 were "unacceptably" wet inspite of surgical therapy and 2 were re-

diverted. Mundy,[123] in a later study, reported that only 3 of 31 were incontinent; however, 21 of these patients received artificial sphincters, and 4 were augmented. Thus, while evaluating the incontinence rate of any study one must critically determine how the patients were initially handled and how the author defines incontinence.

The management of persistent incontinence consists of anticholinergic therapy if one suspects bladder instability as the etiology.[126] In patients who have augmentation cystoplasty with a segment of bowel, dicyclomine is helpful in blunting intestinal peristaltic contractions which may be severe enough to intermittently increase intraluminal pressures above the leak point pressure. Ephedrine, an alpha-adrenergic agonist, can increase bladder neck tone and in a few cases may promote continence even when used as a single agent. Oxybutinin is also efficacious in these cases.[184] Other factors to consider when evaluating results include constipation and concurrent respiratory problems.[182]

Renal Deterioration

Although worsening renal function is often an indication for undiversion, it can also be a complication. Hendren[181] acknowledges that renal dysplasia is part of the pathology in patients with prune-belly syndrome and that, in some, renal failure is unavoidable. In Hendren's series from 1990[127] 23 of 177 patients required transplantation and an additional 40 were expected to need a renal transplantation within 10 years. Nephrectomy, as part of the reconstruction, is recommended if function is documented to be less than 10% on a differential renal scan. Firlit[130] notes that patients who progress to end-stage renal disease often have poor function prior to undiversion. Overall, renal deterioration requiring revision or even re-diversion is rare and the vast majority of patients are found to have either stable or improved renal function after undiversion (96% in Table 2). In our opinion, even if a patient with poor renal function eventually is transplanted, undiversion has served its purpose if it postpones transplantation. Furthermore the chances of a successful transplant are improved if the bladder or neobladder is cycled prior to transplantation.

Complications Due to G.I. Segment

Metabolic disturbances and mechanical complications due to intestinal segments and problems caused by removal of these segments from intestinal continuity were discussed in detail earlier and will not be repeated.

Gastric segments were devised in an effort to avoid these complications but they produce complications of their own. Removing the gastric patch from the stomach reduces its capacity for a short period of time but usually it is not a long-term problem. Metabolic alkalosis due to loss of hydrochloric acid, sodium and potassium can occur, especially with severe vomiting or diarrhea and salt losing kidneys. This is generally managed with intravenous saline infusions.

Hypergastrinemia can also result from decrease in feedback inhibition of the gastric antrum. This phenomenon may cause excessive salt loss from the gastric patch. If hyponatremia, hypokalemia and metabolic alkalosis occur in conjunction with increased serum gastrin levels, initial management consists of increasing oral acid intake with carbonated drinks (carbonic acid) and ascorbic acid supplements at meal times. This may compensate for diminished intrinsic gastric acid production and reduce hypergastrinemia while preventing dehydration. If medical management is unsuccessful the gastric patch may have to be removed from the bladder.[75]

The most common major complication of gastric segments is the hematuria-dysuria syndrome (HDS), which occurs as a result of gastric mucosal secretory activity in 36% of children.[75] It is believed that urine of patients with diminished renal function loses its buffering capacity. However, some patients have reported symptoms even when their urine pH was normal. If gastrocystoplasty is done on a patient who is anuric or oliguric because of end-stage renal disease or prior diversion, the augmented bladder must be irrigated regularly with saline or bicarbonate. Patients, particularly boys with sensate urethras and adults, present with intermittent dysuria of varying severity, along with gross hematuria, which may also vary in severity. The problem is seen more often if there is any degree of incontinence and patients notice it when they void. Hydrogen ion blockers, bicarbonate bladder irrigation and oral bicarbonates are administered when needed. Long-term use of omeprazole (proton pump inhibitor) is also safe. While the majority of patients require short term or no treatment, one in five has to be placed on chronic therapy.

Stone Formation

Bladder calculi occur in about 30%-52% of patients following bladder augmentation with a similar recurrence rate.[110] For reasons that are not clear, girls seem to be at slightly higher risk than boys. Stones are usually struvite

in composition, but apatite stones have also been reported.[185] Risk factors for formation of calculi include cloacal malformations, vaginal reconstructions, anal atresia, bladder neck reconstruction, and presence of a continent stoma. The risk can be attributed, in most cases, to difficulty in catheterization with subsequent stasis of urine. Recurrent, symptomatic urinary tract infections are another risk factor for obvious reasons. Production of thick mucous by bowel segments may also be a risk factor since stones do not form when gastric patches are used.

Bladder Perforation

Perforation can be an early or late complication with a reported incidence of about 4%-12%.[186,127] Perforation is believed to be the result of chronic ischemia due to overdistention and it occurs at or near the anastomotic site. It is critical for patients with bladder augmentation to catheterize on a regular basis, at 3 hourly intervals initially, gradually increasing to every 4-5 hours depending on the total urine output. Symptoms of perforation include abdominal distention, pain, nausea, vomiting, and fever. One should have a high index of suspicion for bladder perforation, especially in myelodysplastic patients who may have diminished sensation to peritoneal irritation. If a patient at risk presents with these symptoms, an immediate CT cystogram should be obtained for diagnosis followed by prompt operative repair.

Reflux Mild reflux demonstrated on preoperative voiding cystography will, more often than not, resolve spontaneously after restoration of ureteral continuity.[135] It has been postulated that absence of flow into the distal ureters of the diverted urinary tract may be responsible for this phenomenon.[187] Nevertheless, persistent reflux requiring reoperation does occur in about 10%-20% of cases. Hendren[127] reported an 8% reoperation rate for reflux in a series of diversions for megaureter. Bauer[128] found persistent reflux in 2 out of 10 myelomeningocele patients, both of whom had bilateral reflux prior to undiversion. In patients with persistent reflux the urodynamic study should be repeated to identify a missed poorly compliant bladder prior to further corrective surgery.

Conclusions

Undiversion of the previously diverted urinary tract is uncommon now since many diverted patients were reconstructed in the late 1970s and 1980s. A tremendous amount of information was gained at the time.[118] Increasing knowledge of bowel physiology and the development of antireflux and continence procedures has made primary reconstruction the standard of care for children with severe urinary tract dysfunction.[140] Much of what has been learned over the years still holds true today and will continue to be applicable in future cases of urinary tract reconstruction.[188]

In summary, there are numerous options available to the surgeon and careful and thoughtful evaluation and planning is essential to identify the best option for a particular child. A successful outcome is as dependent on patient selection as it is on surgical technique. In addition, the surgeon must be capable and willing to deviate from his preoperative plans if operative findings dictate.

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